

Publications by ISPGHAN members in Pubmed Indexed Journals (March 2021 – June 2021)

MARCH 2021

[1.] Bhaswati C Acharyya, Chandrayee Bhattacharyya, Meghdeep Mukhopadhyay and Saumyabrata Acharyya. Polyethylene Glycol Plus Electrolytes with Stimulant Laxative in Paediatric Faecal Disimpaction: A Randomised Controlled Study. *Pediatr Gastroenterol Hepatol Nutr.* 2021 Mar;24(2):230-237.

A randomised prospective study was undertaken, in a tertiary paediatric gastroenterology centre to find out the outcome of a 2-day disimpaction when a stimulant laxative sodium picosulphate was added to PEG3350+E (PEG+E+PS group) and comparing it with the outcome using PEG3350+E (PEG+E group) alone. Hundred and one children were randomised into two groups to receive PEG+E+PS and PEG+E. Results revealed that PEG+E+PS group proved significantly superior to PEG+E group in most of the efficacy-parameters in terms of disimpaction as well as long-term management of constipation.

[2.] Mohan N, Raghunathan V, Dhaliwal MS, Bhangui P, Tiwari A, Soin AS. Pediatric ABO-incompatible Living Related Donor Liver Transplantation: Experience from Indian Subcontinent. *Indian Pediatr.* 2021 Mar 15; 58(3):281-282.

In this research letter authors have presented their centre experience with pediatric ABO-incompatible liver transplantation between January, 2011 and November, 2018. Out of 203 patients that underwent liver transplant during this period, 8 underwent ABO-incompatible liver transplantation; 4 (3 boys) had blood group O+ve. Median (range) age was 28 (7-91) mo, PELD score was 24.5 (14-42), and pre-transplant antibody titer range was 1:32-1024. Number of plasmapheresis sessions required ranged from 1-6. Post-operatively two patients had rise in antibody titer >64 requiring plasmapheresis. All 8 patients survived without rejection/biliary issues. Mean (range) of post-transplant hospital stay was 19.1 (13-22) d and follow-up period was 38.1 (7.1-84.4) mo. They have concluded that pediatric ABO-incompatible liver transplantation can be successfully performed using plasmapheresis with optimal immune-suppression and vigilant post-op monitoring.

[3.] Mohanty N, Thapa BR, Mathai J, Pai U, Mohanty N, Biradar V, Jog P, Prabhu P. Low Osmolarity Oral Rehydration Salt Solution (LORS) in Management of Dehydration in Children. *Indian Pediatr.* 2021 Mar 15;58(3):266-272. doi: 10.1007/s13312-021-2168-8.

The Indian Academy of Pediatrics constituted a panel of experts from the fields of pediatrics, pediatric gastroenterology and nutrition to update on management of dehydration in children with particular reference to low osmolarity oral rehydration salt solution (LORS) and issue a current practice guideline. It was stressed that advantages of LORS far

out-weigh its limitations. Increased use of LORS can only be achieved by promoting better awareness among public and health-care providers across all systems of medicine. LORS can also be useful in managing dehydration in non-diarrheal illness. More research is required to modify ORS further to make it safe and effective in neonates, severe acute malnutrition, renal failure, cardiac and other co-morbidities. There is an urgent need to discourage production and marketing all forms of ORS not in conformity with WHO approved LORS, under a slogan "One India, one ORS".

[4.] Nagendra Kumar, Ujjal Poddar, Rajnikant Yadav, Hira Lal, Krushna Pani, Surender Kumar Yachha, Anshu Srivastava, and Rakesh Pandey. Autoimmune Sclerosing Cholangitis in Children: A Prospective Case-Control Study. *Pediatr Gastroenterol Hepatol Nutr.* 2021 Mar; 24(2):154-163

Aims of this study were to find the prevalence of autoimmune sclerosing cholangitis (ASC) by using magnetic resonance cholangiography (MRC) in autoimmune hepatitis (AIH) and in non-AIH cirrhosis and to compare clinical presentation and outcome of AIH and ASC. In conclusion, definite feature of ASC is uncommon in children with AIH and seen in just 10.5% of cases. Authors suggest MRC in a select group of children with cholestatic features, IBD and in those who showed poor response to immunosuppression, instead of all children with AIH.

[5.] van Wessel DBE, Thompson RJ, Gonzales E, Jankowska I, Shneider BL, Sokal E, Grammatikopoulos T, Kadaristiana A, Jacquemin E, Spraul A, Lipiński P, Czubkowski P, Rock N, Shagrani M, Broering D, Algoufi T, Mazhar N, Nicastro E, Kelly D, Nebbia G, Arnell H, Fischler B, Hulscher JBF, Serranti D, Arikan C, Debray D, Lacaille F, Goncalves C, Hierro L, Muñoz Bartolo G, Mozer-Glassberg Y, Azaz A, Breclj J, Dezsófi A, Luigi Calvo P, Krebs-Schmitt D, Hartleif S, van der Woerd WL, Wang JS, Li LT, Durmaz Ö, Kerak N, Hørby Jørgensen M, Fischer R, Jimenez-Rivera C, Alam S, Cananzi M, Laverdure N, Ferreira CT, Ordóñez F, Wang H, Sency V, Kim KM, Chen HL, Carvalho E, Fabre A, Quintero Bernabeu J, Alonso EM, Sokol RJ, Suchy FJ, Loomes KM, McKiernan PJ, Rosenthal P, Turmelle Y, Rao GS, Horslen S, Kamath BM, Rogalidou M, Karnsakul WW, Hansen B, Verkade HJ. Impact of Genotype, Serum Bile Acids, and Surgical Biliary Diversion on Native Liver Survival in FIC1 Deficiency. *Hepatology.* 2021 Mar 5. doi: 10.1002/hep.31787. Epub ahead of print.

This multicenter, international combined retrospective and prospective study aimed to provide novel insights by using the largest genetically defined cohort of FIC1 deficiency to study natural history, effects of predicted protein truncating mutations (PPTMs), and possible associations of serum bile acid (sBA) concentrations and surgical biliary diversion (SBD) with long-term outcome. It was concluded that less than half of FIC1 deficiency patients reach adulthood

with native liver. The number of PPTMs did not associate with the natural history or prognosis of FIC1 deficiency. sBA concentrations at initial presentation and after SBD provide limited prognostic information on long-term native liver survival. [6.] Ramavath K, Lal SB, Behera A, Kaman L, Dahiya D, Tandup C, Singh K, Abhinay R, Ganesan D, Venkatesh V. **Necrotizing Enterocolitis Following Pediatric Living Donor Liver Transplant.** *Exp Clin Transplant.* 2021 Apr; 19(4):390-392. doi: 10.6002/ect.2020.0287. Epub 2021 Mar 16.

Cow's milk allergy association with necrotizing enterocolitis (NEC) has not been well determined, and the pathophysiology is still not clear. NEC is very rare following living donor liver transplant. In this case, authors have reported on a 6-year-old boy who was doing well in the postoperative period developed sudden worsening of general condition after he was started on milk feed. On evaluation and reexploration, he was diagnosed with necrotizing enterocolitis and later succumbed to death.

[7.] Naresh Shanmugam, Jagadeesh Menon, Mukul Vij, Ashwin Rammohan, Rajesh Rajalingam, Mohamed Rela. **Total Internal Biliary Diversion for Post-Liver Transplant PFIC-1-Related Allograft Injury.** *Journal of clinical and experimental hepatology.* 2021 Mar 30. doi.org/10.1016/j.jceh.2021.03.008 Epub ahead of print.

Authors have reported the case of a seven-year-old boy who underwent liver transplantation (LT) at the age of 2 years for PFIC type 1. Over the next five years, he developed refractory diarrhea, emaciation, worsening liver function, and steatohepatitis. Aiming to interrupt the enterohepatic circulation, at the age of 7 years, he underwent a total internal biliary diversion. The patient's postprocedure period was uneventful. His diarrhea settled and the transaminases normalized his follow-up liver biopsy after a year showed a complete resolution of steatohepatitis. At 18 months' follow-up, he has gained weight and remains asymptomatic. In this report, authors have highlighted that post-LT complications especially allograft injury related to the pathology of PFIC-1 can be safely and effectively managed by performing a total internal biliary diversion.

[8.] Kumar A, Saxena AK, Bhatia A, Lal S, Rana P, Bawa M, Sodhi KS. **Comparison of unenhanced and contrast-enhanced 3 T magnetic resonance portovenography in children with extra hepatic portal venous obstruction.** *Abdom Radiol (NY).* 2021 Mar 30. doi: 10.1007/s00261-021-03064-1. Epub ahead of print.

This study aimed to evaluate role of 3-T magnetic resonance portovenography (MRPV) in children with extra hepatic portal venous obstruction (EHPVO) and compare unenhanced and contrast-enhanced sequences. Authors have concluded that balanced turbo field echo (BTFE) sequence is the single best unenhanced MR pulse sequence to detect all the vascular structures in children with EHPVO. CE-MRI is not superior to BTFE sequence and should be used at the discretion of the radiologist.

APRIL 2021

[9.] Nirmala Dheivamani, Winston Thomas, Rohit Banerjee, Mallar Mukherjee, Monjori Mitra. **Efficacy of polyethylene glycol 3350 as compared to lactulose in treatment of ROME IV criteria-defined pediatric functional constipation: A randomized controlled trial.** *Indian*

J Gastroenterol. 2021 Apr; 40(2):227-233. doi: 10.1007/s12664-021-01148-w. Epub 2021 Apr 8.

The aim of the study was to compare the efficacy of polyethylene glycol (PEG) 3350 and lactulose in the treatment of pediatric functional constipation. This study proved that the PEG 3350 treatment group had early symptom relief and significant improvement in all the ROME IV-defined criteria compared to the lactulose group in pediatric functional constipation.

[10.] Bandyopadhyay T, Deswal S, Maria A, Phulwara RH, Das P, Ahuja A. **Microvillous inclusion disease as a cause of severe congenital diarrhea in a newborn.** *J Matern Fetal Neonatal Med.* 2021 Apr 11:1-3. doi: 10.1080/14767058.2021.1910656. Epub ahead of print.

In this case report have reported a case of intractable diarrhea in a preterm neonate with Microvillous inclusion disease (MVID) phenotype presented on second day of life with intractable diarrhea. The diagnosis was established by classical electron microscopic findings in the intestinal biopsy sample.

MAY 2021

[11.] Manoj Madhusudan, Srinivas Sankaranarayanan, Ramkumar Ramamoorthy, Deenadayalan Munirathnam and Meena Sivasankaran. **Prolidase Deficiency in Very Early Onset Inflammatory Bowel Disease (VEO-IBD).** *The Indian Journal of Pediatrics (May 2021) 88(5):503.*

In this letter to the editor, authors have reported a 4-y-old boy 2nd born to 3rd degree consanguineous marriage who presented with failure to thrive, developmental delay and recurrent episodes of non-bloody diarrhea since 1 y of age. He had multiple episodes of ear discharge and recurrent skin ulcers involving the lower limbs. Colonoscopy revealed complete loss of normal vascular pattern with superficial erosions of the entire colon and biopsies revealed a crypt destructive colitis which raised the possibility of very early onset inflammatory bowel disease (VEO-IBD). He was started on steroids and his IBD underwent clinical remission. In view of parental consanguinity and clinical presentation in a child with VEO-IBD, next generation sequencing was done which revealed a novel homozygous mutation in Exon 12 of PEPD gene [c.825delC (p.Phe275LeufsTer46)], which was pathogenic for prolidase deficiency (PD). This case is presented for the extreme rarity (Incidence 1–2/1,000,000 persons), and to demonstrate the role of molecular genetics in children with VEO-IBD.

[12.] Lourembam, Radhapyari ; Malik, Rohan ; Bolia, Rishi. **Combined Mutations of Canalicular Transporter Proteins Causing Low Phospholipid-Associated Cholelithiasis and Transient Neonatal Cholestasis in an Infant,** *JPGN Reports: May 2021 - Volume 2 - Issue 2 - p e080 doi: 10.1097/PG9.000000000000080*

Authors have reported an infant with transient neonatal cholestasis (TNC) and low phospholipid-associated cholelithiasis (LPAC) with heterozygous mutations in ABCB4 and ABCB11 who responded to UDCA therapy. Apart from reporting LPAC in infancy, this case report also highlights the complex nature of the genetics of cholestatic disorders.

[13.] Lal SB, Venkatesh V, Kumar A, Anushree N, Seetharaman K, Aneja A, Chaluvashetty SB, Sehgal R. **Liver Abscess in Children-experience From a Single Tertiary Care Center of North India: Etiology, Clinical Profile and Predictors of Complications.** *Pediatr Infect Dis J.* 2021 May 1;

40(5):e179-e184. doi: 10.1097/INF.0000000000003053. PMID: 33847292.

Authors aimed to analyze the clinical profile, etiology, risk factors for complications, management and outcomes of liver abscess (LA) in children. They have concluded that Amebic LA is the commonest cause of pediatric LA in endemic regions and is difficult to differentiate from Pyogenic LA clinically. Percutaneous catheter drainage is safe and effective modality for the management of LA in children. A higher alanine transaminase, prolonged prothrombin time/international normalized ratio and low serum albumin levels (<3 g/dL) at presentation identify complicated LA.

[14.] Vijay P, Lal BB, Sood V, Khanna R, Alam S. **Cystatin C: best biomarker for acute kidney injury and estimation of glomerular filtration rate in childhood cirrhosis.** *Eur J Pediatr.* 2021 May 12. doi: 10.1007/s00431-021-04076-1. Online ahead of print.

The objective of the study was to evaluate the diagnostic and prognostic role of serum cystatin C, urinary neutrophil gelatinase-associated lipocalin (NGAL), and renal resistive index (RRI) in AKI among pediatric cirrhotics (children under 18 years of age). The study concluded that serum cystatin C is a useful biomarker to identify acute kidney injury in cirrhotic children with decompensation. Glomerular filtration rate calculation is more accurate by cystatin-based equations than creatinine-based equations.

[15.] Pawaria A, Khanna R, Sood V, Siloliya M, Benjamin JJ, Kumar G, Alam S. **Subjective global nutritional assessment as a nutritional tool in childhood chronic liver disease.** *Br J Nutr.* 2021 May 14;1-10. doi: 10.1017/S0007114521001604. Online ahead of print.

Objective of this study was to assess subjective global nutritional assessment (SGNA) in children with chronic liver diseases (CLD). Agreement between SGNA and anthropometric measures, prediction of morbidity and death or liver transplantation (LT) at 1-year post-enrolment by SGNA and inter-observer reliability of SGNA were assessed. It was concluded that inter-observer agreement in assessment of SGNA was good (90.2 %). SGNA, in contrast to anthropometric measures, was a better nutritional assessment tool. It is reliable, comprehensive and predicts poor outcome in childhood CLD.

[16.] Lehan E, Wang T, Soboleski D, Acker A, Kehar M. **Inflammatory Bowel Disease and Primary Sclerosing Cholangitis in a Pediatric Patient With Neurofibromatosis Type 1.** *ACG Case Rep J.* 2021 May 14;8(5):e00605. doi: 10.14309/crj.0000000000000605.

The literature available on the association of neurofibromatosis type 1 with inflammatory bowel disease is limited to 7 clinical case reports, and none had comorbid primary sclerosing cholangitis. Authors have described a case of a 15-year-old adolescent boy with neurofibromatosis type 1 who presented with inflammatory bowel disease and primary sclerosing cholangitis.

[17.] Bolia R, Sarma MS, Biradar V, Sathiyasekaran M, Srivastava A. **Current practices in the management of corrosive ingestion in children: A questionnaire-based survey and recommendations.** *Indian J Gastroenterol.* 2021 Jun; 40(3):316-325. doi: 10.1007/s12664-021-01153-z. Epub 2021 May 15.

This survey aimed to understand practices adopted by gastroenterologists, identify lacunae in evaluation and management and suggest a practical algorithm. It was derived that

corrosive ingestion mostly affects 2-5-year olds and is accidental in majority. It can be potentially prevented by proper storage and labelling of corrosives. An algorithm for management was proposed.

[18.] Bolia R, Dhanesh Goel A, Badkur M, Jain V. **Gastrointestinal Manifestations of Pediatric Coronavirus Disease and Their Relationship with a Severe Clinical Course: A Systematic Review and Meta-analysis.** *J Trop Pediatr.* 2021 May 17;67(2):fmab051. doi: 10.1093/tropej/fmab051.

The objectives of this systematic review were to determine the gastrointestinal (GI) manifestations of pediatric COVID-19 and to evaluate their role as risk factors for a severe clinical course. Authors have concluded that diarrhea, nausea/vomiting or abdominal pain are present in nearly one-fifth of all children with COVID-19 through a systematic search of which 55 studies (4369 patients) published before 31 December 2020 with information about the GI manifestations of pediatric COVID-19. The presence of diarrhea portends a severe clinical course.

[19.] Gopalkrishna V, Joshi MS, Chavan NA, Shinde MS, Walimbe AM, Sawant PM, Kalrao VR, Dhongade RK, Bavdekar AR. **Prevalence and genetic diversity of gastroenteritis viruses in hospitalized children < 5 years of age in Maharashtra state, Western India, 2017-2019.** *J Med Virol.* 2021 Aug;93(8):4805-4816. doi: 10.1002/jmv.27085. Epub 2021 May 24.

The majority (79%) of the children were <2 years of age in this study. The prevalence of Rotavirus A (RVA) was 30.5% followed by 14.3% for norovirus, 8.4% for adenovirus, and 5.5% for astrovirus. The severity of the disease was highest in patients with coinfections compared with the patients with a single infection or negative for all. The authors have concluded that after the recent inclusion of rotavirus vaccines as a part of the National Immunization schedule in India, conducting extensive AGE surveillance in children should include nonrotaviruses such as norovirus.

[20.] McFarland, Lynne V.; Srinivasan, Ramesh P; Setty, Rajendra P; Ganapathy, Sridhar; Bavdekar, Ashish; Mitra, Monjori; Raju, Bhaskar; Mohan, Neelam.

Specific Probiotics for the Treatment of Pediatric Acute Gastroenteritis in India: A Systematic Review and Meta-Analysis, JPGN Reports: August 2021 - Volume 2 - Issue 3 - p e079 doi: 10.1097/PG9.0000000000000079. Published on line on 2021 May 27.

The authors have concluded that in India, 2 types of probiotics (*S. boulardii* CNCM I-745 and *L. rhamnosus* GG) significantly shortened both the duration of diarrhea and hospitalization stays in pediatric patients with pediatric acute gastroenteritis.

JUNE 2021

[21.] Menon J, Miraje B, Patel K, Vij M, Hakeem A, Deva- rajan V, Shanmugam N, Srinivas Reddy M, Rela M. **Primary tuberculosis of the graft masquerading pyogenic liver abscess in a pediatric liver recipient.** *Transpl Infect Dis.* 2021 Jun;23(3):e13533. doi: 10.1111/tid.13533. Epub 2020 Dec 6.

Primary tuberculosis (TB) of the graft presenting as multiple liver abscesses is previously unreported. In this case report authors have reported that a 14-month-old male child in the early post liver transplant (LT) period presented with high-grade fever spikes and on evaluation was found to have

multiple pyogenic liver abscesses (PLA) in the CT abdomen. His fever was not responding to intravenous antibiotics and liver biopsy was done which showed numerous acid fast bacilli. Genetic analysis confirmed the bacilli as *Mycobacterium tuberculosis* (MTB). Timely diagnosis and prompt introduction of antituberculosis therapy were lifesaving.

[22.] **Kechar M, Ebel NH, Ng VL, Baquero JER, Leung DH, Slowik V, Ovchinsky N, Shah AA, Arnon R, Miloh T, Gupta N, Mohammad S, Kogan-Liberman D, Squires JE, Sanchez MC, Hildreth A, Book L, Chu C, Alrabadi L, Azzam R, Chepuri B, Elisofon S, Falik R, Gallagher L, Kader H, Mogul D, Mujawar Q, Namjoshi SS, Valentino PL, Vitola B, Waheed N, Zheng MH, Lobritto S, Martinez M. Severe Acute Respiratory Syndrome Coronavirus-2 Infection in Children With Liver Transplant and Native Liver Disease: An International Observational Registry Study. *J Pediatr Gastroenterol Nutr.* 2021 Jun 1;72(6):807-814. doi: 10.1097/MPG.0000000000003077.**

In this multicenter observational cohort study, authors have collected data from 91 patients <21 years (LD 44, LT 47) with laboratory-confirmed SARS-CoV2 infection between April 21 and September 17, 2020 and aim to report outcomes for children with native liver disease (LD) and liver transplant (LT) enrolled in the NASPGHAN/SPLIT SARS-CoV2 registry. They have concluded that although not directly comparable, LT recipients had lower odds of severe SARS-CoV2 infection (vs LD), despite immunosuppression burden. NAFLD patients reported to the registry had higher odds of severe SARS-CoV2 disease. Future controlled studies are needed to evaluate effective treatments and further stratify LD and LT patients with SARS-CoV2 infection.

[23.] **Menon J, Vij M, Sachan D, Rammohan A, Shanmugam N, Kaliamoorthy I, Rela M. Pediatric metabolic liver diseases: Evolving role of liver transplantation. *World J Transplant.* 2021 Jun 18;11(6):161-179. doi: 10.5500/wjt.v11.i6.161.**

The authors have provided a comprehensive review on the role of liver transplantation in the management of metabolic liver diseases in children.

[24.] **Poddar U, Yachha SK, Upadhyaya VD, Kumar B, Borkar V, Malik R, Srivastava A. Endoscopic cystogastrostomy: Still a viable option in children with symptomatic pancreatic fluid collection. *Pancreatol.* 2021 Jun; 21(4):812-818. doi: 10.1016/j.pan.2021.02.004.**

Authors have analyzed their centre experience of 33 endoscopic cystogastrostomies done in 29 children (< 18 years) to find its efficacy and safety. They observed that procedure was successful in 29 of 31 (93.5%) children with no mortality. Adverse events happened in four cases (12.9%). Stents were removed in 26 (90%) after 12 (7-20) weeks and got spontaneously migrated out in 3 (10%) cases. Over a median follow-up

of 26 (5-48) months, 26 (90%) had no recurrence of pseudocyst and 3 (10%) had recurrence of a small, asymptomatic pseudocyst. They have concluded that endoscopic cystogastrostomy is a safe and effective method of draining bulging PFCs in children. The procedure carries acceptable morbidity with minimal recurrence.

[25.] **Nabi Z, Lakhtakia S, Chavan R, Asif S, Basha J, Gupta R, Yarlagadda R, Reddy PM, Kalapala R, Reddy DN. Diagnostic utility of EUS-guided tissue acquisition in children: A tertiary care center experience. *Endosc Ultrasound.* 2021 Jun 26. doi: 10.4103/EUS-D-20-00203. Epub ahead of print.**

In this study, authors have aimed to evaluate the feasibility, safety, and diagnostic utility of endoscopic ultrasound-guided fine-needle aspiration or biopsy (EUS-FNA/FNB) in children (<18 years) with various gastrointestinal diseases. EUS-FNA and-FNB were performed in 42 and 25 children, respectively during the study period. All the procedures could be successfully performed and there was no major procedure-related adverse event. Minor adverse events included self-limiting throat pain (10) and abdominal pain (3), self-limited bleeding at puncture site (3), and transient fever (1). EUS-FNA/FNB provided a histopathological diagnosis in 59 (88.1%) children.

[26.] **Mohan N, Matthai J, Bolia R, Agarwal J, Shrivastava R, Borkar VV; For Pediatric Gastroenterology Chapter Of Indian Academy Of Pediatrics, Indian Society Of Pediatric Gastroenterology, Hepatology And Nutrition (ISPGHAN). Diagnosis and Management of Gastro-esophageal Reflux Disease in Children: Recommendations of Pediatric Gastroenterology Chapter of Indian Academy of Pediatrics, Indian Society of Pediatric Gastroenterology, Hepatology and Nutrition (ISPGHAN). *Indian Pediatr.* 2021 Jun 28;S097475591600348. Epub ahead of print.**

The salient recommendations on gastro-esophageal reflux disease (GERD) in children proposed by the expert group are (1.) careful history and clinical examination are adequate to make a diagnosis in most patients, but judicious investigations are necessary in a few (2.) upper gastro intestinal tract endoscopy may be required in those with esophageal manifestations, dysphagia and hematemesis (3.) in children with extra-esophageal symptoms, multichannel intraluminal impedance (MII-Ph) monitoring and scintigraphy are necessary (4.) Empirical treatment with a Proton pump inhibitor (PPI) has not been proven useful in infants, but a four-week trial is recommended in older children without complications (5.) While positioning and feed thickening have limited benefit in infants, life-style modifications are important in older children. Separate algorithms for management of infants and older children with suspected GERD have also been proposed.

Compiled by Dr.Prasanth.K.S